Diagnosis of deafness in a horse by brainstem auditory evoked potential

Malte M. Harland, Allison J. Stewart, Arvle E. Marshall, Ellen B. Belknap

Abstract — Deafness was confirmed in a blue-eyed, 3-year-old, overo paint horse by brainstem auditory evoked potential. Congenital inherited deafness associated with lack of facial pigmentation was suspected. Assessment of hearing should be considered, especially in paint horses, at the time of pre-purchase examination. Brainstem auditory evoked potential assessment is well tolerated and accurate.

Résumé — Diagnostic de surdité chez un cheval par potentiel auditif évoqué au niveau du tronc cérébral. Un diagnostic de surdité a été confirmé par potentiel auditif évoqué au niveau du tronc cérébral chez un cheval Paint Horse overo aux yeux bleus âgé de 3 ans. Une surdité congénitale héréditaire associée au manque de pigmentation faciale a été suspectée. Une évaluation de l'audition devrait être envisagée, particulièrement chez les Paint Horses lors de l'examen précédant l'achat. L'évaluation par potentiel auditif évoqué au niveau du tronc cérébral est bien tolérée et précise.

(Traduit par Docteur André Blouin)

Can Vet J 2006;47:151–154

Deafness should be suspected in horses that are unresponsive to external noise. A twitch of the ears or turning of the head in response to a sudden noise, such as clapping the hands, is called the startle reaction and rules out complete deafness. Deaf horses may spook easily, as they are reliant on visual and tactile stimuli. Similar to deaf people, deaf animals rarely have any mental deficits and, with careful training, they can be useful performance horses. However, deafness is a defect and consideration should be given to decreasing the prevalence of deafness in paint horses by selective breeding. An attempt to assess hearing ability should also be made by veterinarians performing prepurchase examinations and by judges at breed shows. Other than anecdotal reports and a personal communication (KD Magdesian, 1998), reports of deafness in paint horses are lacking in the veterinary literature.

The startle test does not assess hearing thresholds, thus it is used as presumptive evidence of deafness and does not distinguish between peripheral and central nervous system defects. Hearing can be definitively assessed by the use of brainstem auditory evoked potential (BAEP) (1). The procedure does not require general anesthesia and is well tolerated by horses, often with no sedation and minimal restraint (2). Headphones are held over, or placed in, the horse's ears and a series of click noises of various intensities are generated to stimulate the auditory pathway from the ear to the brain. In an individual hearing normally, the record of the evoked potential will consist of a series of 5 to 7 characteristic waves, depending on the species (3,4). In animals with congenital deafness, no waves are observed. Both ears should be tested individually to rule out unilateral deafness. Animals that are deaf in 1 ear can be difficult to identify without BAEP.

Department of Clinical Sciences (Harland, Stewart, Belknap) and Department of Anatomy, Physiology and Pharmacology (Marshall), College of Veterinary Medicine, Auburn University, Wire Road, Auburn, Alabama, USA.

Dr. Harland's current address is Nevelstrasse 71, Friedrich-Ebert-Allee 1, D-23879 Moelln, Germany.

Address all correspondence and reprint requests to Dr. A. Stewart; e-mail: stewaaj@vetmed.auburn.edu

Case description

A 3-year-old, gelded, sorrel and white overo American paint horse was examined because of suspected deafness. As a foal, he appeared unresponsive to environmental noises. When used for western pleasure competitions, he appeared oblivious to carnival noise. The owners had never heard the horse vocalize. There was no historical evidence of visual deficits, head trauma, or exposure to ototoxic drugs. A related blue-eyed horse was also suspected to be deaf.

The gelding was in excellent body condition and weighed 501 kg. Hearing ability, as judged subjectively by the startle test, was absent. The right iris was blue, while heterochromia iridis was observed in the left eye with a combination of blue and brown. The remainder of the cranial nerve and physical examinations gave normal results. Results from a complete white blood cell count and serum biochemical panel were within the reference ranges.

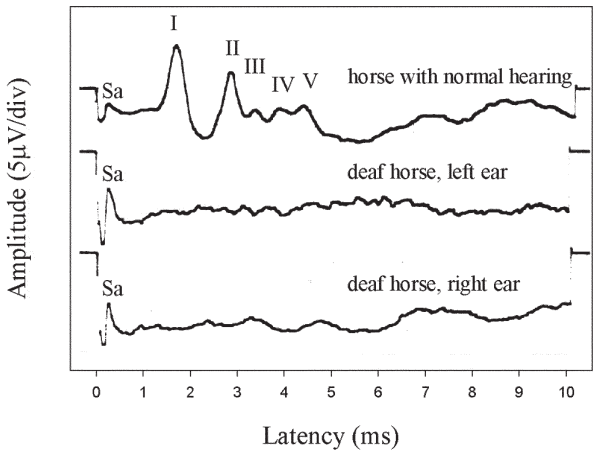

Figure 1. Comparative brainstem auditory evoked potential (BAEP) recordings elicited by air conducted click stimuli on a horse with normal hearing (75 dB, 1750 clicks), and a deaf horse (left ear: 103 dB, 2250 clicks; right ear: 103 dB, 2250 clicks). Sa: Stimulus artefact.

To assess the apparent deafness, the BAEP on the case horse was tested. To validate the test, the BAEP was also assessed on a control horse. Both horses were sedated with detomidine hydrochloride (Dormosedan; Pfizer Animal Health, Exton, Pennsylvania, USA), 0.013 mg/kg bodyweight (BW), IV. Three platinum needle electrodes (Model E2B; Grass Instrument Company, Quincy, Massachusetts, USA) were inserted subdermally on the horses' heads with the reference electrode (+) placed at the vertex and the active recording electrode (-) placed laterally at the most ventral portion of the palpable ear canal of the ear to be stimulated. The ground electrode was inserted at the same site as the active electrode, but on the contralateral ear. A magnetic headphone (Model TDH-39; Nicolet Biomedical Instrument Company, Madison, Wisconsin, USA) was held against the pinna, so that the click sound (stimulus) would be directed down the external ear canal. The headphone was connected to a square wave pulse generator (Model S 10 CTCMA; Grass Instrument Company), which was adjusted to deliver clicks (0.1 ms duration, 103 dB sound pressure level [SPL], [75 dB SPL in the control horse] as measured at the headphone), at a rate of 20 clicks/s. The stimulator triggered the signal averager recording system (Model TD 20; TECA Corporation, Pleasantville, New York, USA). The recording equipment settings were as follows: 1) amplification at display: 5 mV/cm (vertical); 2) sweep rate: 1 ms/division; and 3) band-pass filters: 200 Hz — 2 kHz. The test was repeated a sufficient number of times on the control horse to obtain a clear recording, and on the deaf horse to obtain a quiet background record. Two independent sets of ipsilateral recordings were taken and superimposed to ensure reproducibility. Each ear was tested independently.

The response in the control horse consisted of a series of 5 wave peaks (I-V) (Figure 1). The BAER in the suspected deaf horse showed an absence of all peaks (Figure 1), thus confirming sensorineural deafness, with the problem originating in the inner ear, rather than at some point further along the auditory pathway.

Bone conducted BAEP was attempted in an effort to differentiate between sensorineural and conductive deafness. The bone stimulator was placed on the temporal bone, just dorsal to the temporomandibular articulation, at the buccal surface of the 3rd upper molar by using a mouth speculum, and in other locations, as performed in dogs, where the stimulator is placed on the vertex, midline, just caudal to the interorbital line, and on the zygomatic arch (5). The technique was unsuccessful in obtaining a response in the control horse; therefore, it was pointless to attempt the method in the deaf horse.

Discussion

Congenital deafness associated with pigmentation (white coat or blue eyes) has been reported in several species, including humans, dogs, cats, and mink (6–10). The horse in this report was a blue-eyed, sorrel and white paint horse. Blue eyes in horses occur mainly in splashed white overos and in toveros and bald-faced horses (11). It is a common myth that all splashed white horses are deaf. Suspected congenital deafness in 6 predominantly white American paint horses with either a uni- or a bilateral blue iris was confirmed in 4 cases by BAEP, (personal communication 1998, KD Magdesian).

Deafness in this horse was confirmed by BAEP, and a congenital etiology was suspected based on the owner's verbal history. Histologic studies of the inner ear would be required to definitively prove a congenital defect. Inherited congenital sensorineural deafness, the cause of deafness in most white-coated cat and dog breeds, results from perinatal degeneration or dysfunction of the stria vascularis. The stria vascularis forms the vascular bed of the outer wall of the cochlear duct and is thought to secrete endolymph. Strial deterioration results from the absence or suppression of its melanocyte layer by species specific genes (12), and leads to hair cell deterioration. The same pathophysiology is likely, but has not been proven, in blue-eyed horses lacking pigmentation of the face.

Genetic correlation estimates in other species between deafness, eye color, and hair color patterns reveal strong interrelationships among these characteristics. Investigations of phenotypic markers (sex, hair coat color, pigmentation of different areas of skin, iris and retinal tapetal pigmentation) and sire and dam BAEP status have been performed in dogs, mainly Dalmatians (8,9), and blue-eyed white cats (10).

To investigate the cause of deafness in an individual, a detailed case history (onset of suspected deafness, previous disease, and therapy) is important, as many factors can contribute to deafness. Humans with blue irises are 5.8 times as likely to be deafened by meningitis as those with dark irises (13), and a reversible reduction in hearing after acute salicylate intoxication has been reported (14). Chronic renal failure in children often results in subclinical disturbances in neural conduction of the auditory pathway, which suggests a central component of uremic axonopathy in the pathogenesis of hearing impairment (15). The auditory system is also highly sensitive to bilirubin toxicity (16). Fetal aminoglycoside ototoxicity has been reported in humans (17), but this has not been investigated in foals. However

gentamicin, 5 mg/kg, IM, q8h administered for 7 d in 3 ponies, and for 11 d in 3 other ponies did not affect auditory response, assessed by BAER every 5 d, for up to 60 d after the first dose (18).

Deafness can be classified as central or peripheral. Central deafness is retrocochlear. Peripheral deafness is characterized as inherited or acquired, congenital or late-onset, and sensorineural or conductive. Deafness in horses is rarely evaluated, and to our knowledge, diagnosis of deafness has only been reported once (personal communication 1998, KD Magdesian). The BAEP has been used for diagnosis of deafness in humans, dogs, and cats (7–9), ototoxicity studies in dogs, calves, ponies, and horses (18–20), and inner ear injuries in horses (21).

The brainstem auditory evoked response is a recording of the response of specific structures of the auditory pathway in the caudal brainstem to an appropriate stimulus. The BAEP is classified as a far-field recording, in contrast to a near-field recording in which the recording electrodes are positioned directly into the brainstem. Far-field recording requires a signal averaging method to accentuate the potential waveforms. Stimuli of the BAEP are conducted through either air or bone. Air conducted stimuli are usually earphone produced clicks to the auditory canal that are conducted to the fluids of the inner ear, producing recordable activity in the auditory nerve and brainstem.

Air-conducted click stimuli can be used to diagnose sensorineural deafness, if conductive deafness can be ruled out. Conductive deafness may be due to developmental defects affecting the ossicles or from occlusion of the ear canal (either congenital or secondary to otitis externa or media). Conductive deafness can be diagnosed by recording BAEP responses elicited by a vibrating stimulus transducer held in contact with the skull. Bone conducted stimulation in humans bypasses the outer and, possibly, the middle ear, and directly activates the cochlea, ossicles, or both (22,23). The major acoustic pathway in bone conducted stimulation to the inner ear is through skull contents, probably the cerebrospinal fluid (23,24). The lack of bone conduction BAEP in the control was most likely due to the large size of the horse's head and the relatively small vibrating transducer (Model 934-HSB, TECA Corporation, Pleasantville, New York, USA), causing signal attenuation that precluded the use of this technique to differentiate sensorineural from conductive deafness in the deaf horse.

The wave forms that comprise the BAEP consist of 5 to 7 peaks that occur within 10 msec of the stimulus, as recorded from the surface of the head (3,4). The wave peaks represent activity in the auditory nerve and sequential contributions from several nuclei of the brainstem auditory path. The 5th wave peak is the result of caudal colliculus activity. These wave sources were initially confirmed in studies on cats (25) and by correlating specific lesions in humans with the diagnostic test records (26). By inference, it is presumed that the wave forms correlate with those of all other species that possess a similar mammalian auditory path anatomy. By analyzing the recorded waves in terms of completeness, amplitudes, and latencies, hearing ability is evaluated, and lesions may be localized within the auditory pathway. Midbrain deafness (27), as well as diseases of the vestibular organ and cortical deafness (28), do not influence BAEP. Reference BAEPs have been recorded on horses, ponies, and foals without hearing deficits (29–31). Brainstem auditory evoked potential recording can be performed in anesthetized, sedated, or unsedated horses (1,2,32).

The BAEP appears to be a practical, rapid, well-tolerated, and accurate diagnostic tool in horses with suspected deafness. Evidence from humans suggests that the BAEP might be questionable during the 1st postnatal day, because of conductive hearing loss caused by absorption of amniotic fluid from the middle-ear cavity (33). Nevertheless, accurate BAEP recordings have been performed successfully in 2- to 4-day-old foals (29). By testing the BAEP, deafness can be assessed after the 1st d post-partum in the horse. However, several breeds of dogs (including dalmatians) that have sensorineural congenital deafness are born with the ability to hear, but become deaf by appoximately 6 wk of age, after degeneration of the stria vascularis (34). The defect and time of onset of congenital inherited deafness in paint horses is unknown; therefore, hearing testing in foals may need to be delayed or repeated later in life if deafness is suspected. Although deaf animals generally have normal mental capacities, owners should be made aware that bilaterally deaf horses are reliant on visual and vibratory sensory information, and are therefore more likely to be startled. Assessment of hearing should be considered, especially in paint horses, at the time of prepurchase examination. Lack of a startle response should be further investigated with BAEP.

Acknowledgment

The authors thank Dave Storrs (Licensed Veterinary Technician) for valuable technical help and advice. CVJ

References

1. Rolf SL, Reed SM, Melnick W, Andrews FM. Auditory brain stem response testing in anesthetized horses. Am J Vet Res 1987;48: 910–914.
2. Marshall AE. Brain stem auditory-evoked response in the nonanesthetized horse and pony. Am J Vet Res 1985;46:1445–1450.
3. Davis H. Principles of electric response audiometry. Ann Otol Rhinol Laryngol Suppl 1976;85:1–96.
4. Picton TW, Hink RF. Evoked potentials: How? What? and Why? Am J EEG Technol 1974;14:9–44.
5. Strain GM, Green KD, Twedt AC, Tedford BL. Brain stem auditory evoked potentials from bone stimulation in dogs. Am J Vet Res 1993;54:1817–1821.
6. Brown KS, Bergsma DR, Barrow MV. Animal models of pigment and hearing abnormalities in man. Birth Defects Orig Artic Ser 1971;7:102–107.
7. Famula TR, Oberbauer AM, Williams DC. Gender effects in hearing loss in Dalmatians. Prev Vet Med 2001;48:15–24.
8. Juraschko K, Meyer-Lindenberg A, Nolte I, Distl O. Analysis of systemic effects on congenital sensorineural deafness in German Dalmatian dogs. Vet J 2003;166:164–169.
9. Strain GM. Deafness prevalence and pigmentation gender associations in dog breeds at risk. Vet J 2004;167:23–32.
10. Bergsma DR, Brown KS. White fur, blue eyes, and deafness in the domestic cat. J Heredity 1971;62:171–185.
11. Sponenberg DP. Equine color genetics. Iowa: Iowa State Univ Pr, 2003.
12. Strain GM. Aetiology, prevalence and diagnosis of deafness in dogs and cats. Br Vet J 1996;152:17–35.

13. Cullington HE. Light eye colour linked to deafness after meningitis. BMJ 2001;322:587.
14. Wecker H, Laubert A. Reversible reduction in hearing with acute salicylate intoxication, HNO 2004;52:347–351.
15. Orendorz-Fraczkowska K, Makulska I, Pospiech L, Zwolinska D. The influence of haemodialysis on hearing organ of children with chronic renal failure. Otolaryngol 2002;56:597–602.
16. Shapiro SM, Nakamura H. Bilirubin and the auditory system. J Perinatol 2001;21:52–55,59–62.
17. Niebyl JR. Antibiotics and other anti-infective agents in pregnancy and lactation. Am J Perinatol 2003;20:405–414.
18. Nostrandt AC, Pedersoli WM, Marshall AE, Ravis WR, Robertson BT. Ototoxic potential of gentamicin in ponies. Am J Vet Res 1991;52:494–498.
19. Morgan JL, Coulter DB, Marshall AE. Effects of neomycin on the waveform of auditory-evoked brain stem potentials in dogs. Am J Vet Res 1980;41:1077–1081.
20. Merchant SR, Neer TM, Tedford BL. Ototoxicity assessment of a chlorhexidine otic preparation in dogs. Prog Vet Neurol 1993; 4:72–75.
21. Marshall AE, Byars TD, Whitlock RH, George LW. Brainstem auditory evoked response in the diagnosis of inner ear injury in the horse. J Am Vet Med Ass 1981;178:282–286.
22. Sohmer H, Freeman S. The latency of auditory nerve brainstem evoked responses to air- and bone-conducted stimuli. Hear Res 2001;160:111–113.
23. Kirikae I. An experimental study on the fundamental mechanisms of bone conduction. Acta Otolaryngol Suppl 1959;145:1–145.
24. Sichel JY, Freeman S, Sohmer H. Lateralization during the Weber test: animal experiments. Laryngoscope 2002;112:542–546.
25. Achor LJ, Starr A. Auditory brain stem responses in the cat. 1. Intracranial and extracranial recordings. Electroenceph Clin Neurophysiol 1980;48:154–172.
26. Chiappa KH. Brain Stem Auditory Evoked Potentials: Interpretation. New York: Raven Pr, 1989.
27. Vitte E, Tankere F, Bernat I, Zouaoui A, Lamas G, Soudant J. Midbrain deafness with normal brainstem auditory evoked potentials. Neurology 2002;58:970–973.
28. Delauche AJ. Brain-stem evoked responses as a diagnostic tool for deafness; a neurophysiological test with potential? Br Vet J 1996; 152:13–15.
29. Steiss JE, Bredemuehl JP, Wright JC, Storrs DP. Nerve conduction velocities and brain stem auditory evoked responses in normal neonatal foals, compared to foals exposed to endophyte-infected fescue in utero. Prog Vet Neurol 1991;2:252–260.
30. Mayhew IG, Washbourne JR. Brainstem auditory evoked potentials in horses and ponies. Vet J 1997;153:107–113.
31. Mayhew IG. The clinical utility of brainstem auditory evoked respone testing in horses. Equine Vet Educ 2003;15:27–33.
32. Mayhew IG, Washbourne JR. Short latency auditory evoked potentials recorded from non-anaesthetized thoroughbred horses. Br Vet J 1992;148:315–327.
33. Priner R, Freeman S, Perez R, Sohmer H. The neonate has a temporary conductive hearing loss due to fluid in the middle ear. Audiol Neurootol 2003;8:100–110.
34. Mair IWS. Hereditory deafness in the Dalmation dog. Arch Otorhinolaryngol 1976;212:1–14.